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Case Report

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Occult T_3 Toxicosis in McCune-Albright Syndrome

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Key Words
McCune-Albright syndrome
Thyrotoxicosis

Abstract
We report a girl with McCune-Albright syndrome who presented with Cushing syndrome from adrenal hypersecretion and gonadotrophin-independent precocious puberty in the first year of life. At age 5, she failed to gain weight and was found to have hyperthyroidism, which was occult in that she had T_3 toxicosis without a goitre or thyroid ultrasound abnormality. The latter has not been previously reported in McCune-Albright syndrome.

Introduction

McCune-Albright syndrome is caused by post-zygotic somatic mutations leading to enhanced function of G-α-S protein [1] and is associated with multiple endocrinopathies, such as adrenal hyperplasia, gonadotrophin-independent precocious puberty, pituitary hypersecretion, hyperparathyroidism and hyperthyroidism.

Case Report

A 5-year-old girl with McCune-Albright syndrome was investigated for failure to thrive. She was born by emergency caesarean section for fetal distress at 37 weeks gestation weighing 1.6 kg. At the age of 7 weeks, she was admitted with bilious vomiting and abdominal distention and, at laparotomy, bilateral multiloculated cysts from the ovaries were drained. At the age of 6 months, she was re-admitted because of weight loss and diarrhoea. She was noted to be cushingoid and had several large irregular-edged café-au-lait patches on her trunk and sacrum which had not been present in the neonatal period. Plasma cortisol levels were markedly elevated with levels of 1,462 and 1,420 nmol/l at 09.00 and 24.00 h. Her ACTH level was low at less than 7 ng/l. Plasma cortisol levels failed to be suppressed by high-dose dexamethasone. She underwent bilateral adrenalectomy at the age of 9 months and the adrenals were demonstrated to have nodular hyperplasia on histological examination. She was treated with replacement glucocorticoid and mineralocorticoid. She continued to have gonadotropin-independent preococius puberty, which was treated with cyproterone acetate. She developed a pathological fracture of the femur from polyostotic fibrous dysplasia which failed to unite. She continued to have severe failure to thrive and developmental delay. Of note, repeated thyroid function tests revealed a normal plasma free T_4 concentration.

At the age of 5, she was failing to gain weight, despite a substantial calorie intake of 145 kcal/kg/24 h (average for age, 80–90) [2]. She was not sweaty, tachycardic or irritable and there was no history of diarrhoea. There was no goitre. Her growth velocity over the previous 2 years had been 2 cm per year (–4.3 SDS). The only parameter to suggest thyrotoxicosis was a high energy requirement, despite being wheelchair-bound due to her orthopaedic difficulties. Free T_4 was 16.0 nmol/l (NR, 9.0–23.8) and free T_3 10.0 pmol/l (NR, 2.5–5.3). Thyrotrophin-releasing hormone stimulation test failed to stimulate TSH secretion (TSH less than 0.04, 0.2 and 0.05 mU/l at 0, 20 and 60 min following TRH administration, respectively). Plasma free T_3 was assayed by a microparticle enzyme-linked immunoassay (Abbott Laboratories IMX assay). Unexpectedly, an ultrasound of the thyroid gland was normal.

Following treatment with carbimazole, her weight velocity dramatically increased, despite a decrease in calorie intake. Coincidentally, her growth rate increased to 8 cm per year.

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Discussion

In McCune-Albright syndrome, thyroid dysfunction, like that of the ovaries [3], is associated with structural abnormalities. Elevated free T_3 levels, in combination with suppressed basal and stimulated TSH levels, have been reported only in the presence of ultrasound abnormalities of the thyroid gland in children with this disorder [4]. The ultrasound scan in our patient, however, failed to demonstrate any thyroid abnormality. The clinical feature of thyrotoxicosis was solely of a high energy consumption in the presence of failure to thrive. Thyrotoxicosis was occult both clinically and biochemically and emphasises the importance of measuring free T_3 in addition to other thyroid hormones.

Acknowledgment

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Congress Calendar

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04.09.–07.09.1998 **San Francisco, Calif.** USA	**Growth Hormone Research Society Conference**	**Contact:** J.S. Christiansen, Medical Department M, Aarhus Kommunehospital, DK–8000 Aarhus C, Denmark Tel: +45 86 12 5555 ext 2084; Fax: +45 86 12 5013
16.09.–19.09.1998 **Portland** USA	**71st Annual Meeting of the American Thyroid Association**	**Contact:** Diane P. Miller Tel: +1 718 882 6047; Fax: +1 718 882 6085
24.09.–27.09.1998 **Florence** Italy	**37th ESPE Meeting**	**Contact:** Prof. G. Giovannelli, Istituto di Clinica Pediatrica, Via A. Gramsci, 14, I–43100 Parma, Italy Tel: +39 521 290458; Fax: +39 521 290458
12.04.–16.04.1999 **Bournemouth** UK	**18th Joint Meeting of the British Endocrine Societies**	**Contact:** Amanda Sherwood, Society for Endocrinology, 17/18 The Courtyard, Woodlands, Almondsbury, GB–Bristol BS12 4NQ, UK Tel: +44 1454 619 036; Fax: +44 1454 616 071
30.04.–03.05.1999 **San Francisco, Calif.** USA	**Pediatric Academic Societies (PAS) Annual Meeting**	**Contact:** Registration, 141 Northwest Point Blvd., P.O. Box 675, Elk Grove Village, IL 60009-0675, USA Tel: +1 708 427 1205; Fax: +1 708 427 0275
28.09.–03.10.1999 **Palm Beach, Fla.** USA	**72nd Annual Meeting of the American Thyoid Association**	**Contact:** Diane P. Miller Tel: +1 718 882 6047; Fax: +1 718 882 6085